Combined General Anaesthesia with Retrolaminar block for breast cancer surgery in Pompe’s disease

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Abstract

Pompe’s disease is a rare metabolic disease that results in progressive muscle weakness which culminates in respiratory failure in the late onset type. Patients with Pompe’s disease are at high risk of cardiac and respiratory complications under general anaesthesia owing to their premorbid cardiac and respiratory impairments, as well as the respiratory depressant effects of anaesthetic and analgesic agents. We present a high-risk case of a patient with late onset Pompe’s disease complicated by severe restrictive lung disease scheduled for breast cancer surgery under general anaesthesia with a single shot retrolaminar nerve block for analgesia.

Introduction

Pompe’s disease is a rare genetic condition that presents with progressive weakness of the cardiac and skeletal muscles.¹ Late-onset Pompe’s disease (LOPD) presents with worsening muscle weakness which may progress to respiratory failure but typically spares the heart.¹ Anaesthetic for patients with LOPD is thus complicated by the impairments in diaphragmatic function²,³, respiratory muscle weakness, and elevated perioperative respiratory risks.

Most case reports on anaesthetic management for Pompe’s disease are primarily focused on early-onset Pompe’s disease. Here, we report a case of a 30-year-old patient with LOPD and severe restrictive lung disease for breast cancer surgery with a combined general and regional anaesthetic technique.

Case Report

A 30-year-old female patient with a background of Pompe’s disease presented for left mastectomy, sentinel lymph node biopsy, frozen section and axillary sampling for newly diagnosed stage 2 left breast cancer.

She was diagnosed at age 14 with LOPD and had been receiving regular alglucosidase alfa infusion since the age of 22. Her condition was
complicated by proximal myopathy, respiratory muscle weakness, chronic Type 2 respiratory failure requiring nocturnal Bilevel Positive Airway Pressure device (BiPAP) for the past 7 years, and severe restrictive defect (FEV1 45% predicted). Baseline saturations on room air were 99%, but patient reported breathlessness when bending forward to wear shoes and had difficulty getting back upright if lying down. Functionally, she was otherwise independent in her activities of daily living (ADLs) and was ambulant with crutches in the community.

Preoperatively, the anaesthetic team recommended regional anaesthetic technique for this patient given her severe restrictive lung disease and high pulmonary risk for general anaesthesia. However, the patient requested a general anaesthetic as she was anxious about being awake during the surgery despite the elevated risks. Hence, a combined general anaesthetic and regional technique was used for the surgery.

With the patient awake, a retrolaminar block was performed at the level of T4 with 25ml 0.3% ropivacaine. A dexmedetomidine infusion was started at 0.5mcg/kg/h. The patient was then induced with propofol 1.5mg/kg and fentanyl 1mcg/kg followed by insertion of a laryngeal mask airway (LMA) and maintenance of anaesthesia with sevoflurane. The patient was on Pressure Support ventilation mode with no muscle relaxant administered throughout the operation. At the end of the surgery, Pectoralis II block was done under direct vision by surgeons with 30ml of 0.3% ropivacaine. LMA was removed with tidal volumes on spontaneous ventilation. A total of 137.5mcg of fentanyl and 1g of paracetamol were administered intraoperatively for analgesia.

She was then transferred to the high-dependency ward postoperatively where she was monitored for a day before being stepped down to the general ward.

Discussion

Anaesthesia for patients with LOPD has been reported to be complicated by postoperative respiratory failure, prolonged respiratory depression, and even airway stenosis from worsening of spinal lordosis. As such, significant attention should be paid to the patient’s preoperative respiratory status to identify high-risk patients and provide them with the appropriate preoperative counseling, allow for planning of anaesthetic for risk mitigation, and to determine requirement for postoperative disposition in intensive care as required.

The use of nocturnal BiPAP and functional weakness were signs of her premorbidly compromised respiratory function. This patient’s severe restrictive lung defect is likely related to underlying scoliosis, which has been found in up to 1/3 of patients with Pompe’s disease. The restrictive defect, adding to her pre-existing respiratory weakness, further compounds her already delicate respiratory status.

This operation could have been performed under a purely regional technique with the added benefits of reduced respiratory depression, reduced postoperative nausea and vomiting, better pain relief, shorter hospital stay and reduced time to ambulation. However, owing to the patient’s wishes for a general anaesthetic to ease her anxiety regarding the operation, the team opted for a combined general anaesthetic with a regional technique, avoiding the use of neuromuscular blockade, to reduce her risk of postoperative respiratory failure. A multimodal analgesic technique was also used, including the use of dexmedetomidine infusion, local anaesthetic infiltration and paracetamol to minimize opioid use, and hence reducing the risk of respiratory impairment.

In this patient, we had opted to perform a retrolaminar block, a modification of the paravertebral block aiming for the needle to
contact the retrolaminar site. Local anaesthetic is then injected between the plane of the bone and the overlying paraspinal muscle where it spreads into the paravertebral space.

Use of the retrolaminar catheter technique had previously been described for breast surgery with good analgesic effect, with no requirement for rescue opioids in the postoperative and inpatient period. The main advantage of the retrolaminar block over the paravertebral block is reduced risk of pneumothorax and pleural puncture as the needle is directed medially away from the pleura to contact the bone. Another study reported that the retrolaminar block was quicker to perform compared to the paravertebral block due to easier visualization of target structures and hence easier needle advancement. However, a study on breast surgery patients comparing analgesic requirements with or without retrolaminar block suggested that analgesic requirements were similar postoperatively. Another study suggested that while retrolaminar block was effective in reducing pain score in the immediate post-operative period, its effect was not sustained in the 12h postoperative phase. The conflicting data regarding efficacy of the retrolaminar block for analgesia may be due to variability of spread of local anaesthetic to the paravertebral space as suggested by cadaveric studies. Furthermore, the spread of local anaesthetic may be volume dependent and at present, the recommended volume of injectate in retrolaminar block is still unclear and more studies may need to be performed to establish this.

**Conclusion**

In conclusion, this case describes a successful anaesthetic management of this high-risk patient with Pompe’s disease and pre-existing respiratory impairment. A multimodal analgesic technique combined with regional anaesthesia is important in reducing both opioid and anaesthetic requirements as well as related respiratory depression which may compromise respiratory function leading to postoperative respiratory failure. retrolaminar block is a safe alternative to the paravertebral block in patients at high risk of respiratory compromise.

**References**


